

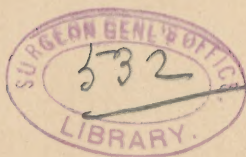
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WITH REPORT OF A CASE.

BY

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THIS rare skin disease was first reported by Addison and Gull in *Guy's Hospital Reports* in 1851. Since then Hutchinson, Bristowe, Gendre (case of Hillairet), Malcolm Morris (two cases), Besnier, Chambard (case of Hillairet), Hardaway, Barlow, Cavafy, Colcott Fox, Vidal, Crocker, Robinson, Payne, and Johnston have published cases. This would make a total of eighteen cases (inclusive of mine) on record at the present time. Aubert related a case to Chambard that had occurred in his practice, but a description of this case has never been published.

It is noteworthy that ten out of the seventeen cases have been reported by English dermatologists. To my knowledge, no case has as yet been reported in Germany or Austria. Malcolm Morris deserves much credit for recognizing this disease as separate and distinct from, yet related to, the ordinary xanthoma. It is not probable that this affection is as rare as the above figures would seem to indicate. Glycosuria exists unsuspected in many cases. As soon as the general practitioner is able to recognize the clinical features of this disease, the number of reported cases will multiply. In quite a number of the cases published the glycosuric condition was only discovered after the character of the eruption had given the clew.

Xanthoma diabeticorum may be described as a subacute inflammatory affection of the skin, associated with glycosuria, and characterized by an eruption of discrete or confluent, firm, dull-red, pinhead to pea sized papules, with often yellowish summits, coming on rather rapidly, and disappearing as the constitutional condition improves. The yellowish "heads" give the lesions a pustular appearance, but upon incision they are found to be solid. This condition is not uniformly



present. Dilated capillaries are often observed over the papules, and produce a hazy redness which tends to obscure their yellow color.

Itching, prickling, or tenderness is usually experienced in the lesions. From a perusal of the cases reported, it would seem that tenderness on pressure is the most constant subjective symptom.

Ætiology.—The disease is nearly always associated with glycosuria. In one case it antedated the appearance of sugar in the urine. It has, however, been absent in a few of the cases. It would seem that the eruption and the glycosuria are both symptoms of an altered hepatic metabolism, and that the one may exist without, of necessity, the other. With but two exceptions the patients have been males, their ages varying from twenty-six to forty-eight. They have nearly all been corpulent and most of them in apparently excellent health.

Prognosis.—The prognosis is favorable. The disease has in some cases evinced a marked tendency to recurrence. Besnier reports a case in which for five consecutive years the eruption made its appearance in the month of July and disappeared in November. Robinson's case also recurred. The French have, therefore, called the disease, the transitory remittent or intermittent xanthoma of diabetics. The last two terms are inaccurate, as the majority of cases do not recur. The disease tends to rapid involution and may disappear in a few weeks, although the average convalescence requires, perhaps, several months.

Diagnosis.—The diagnosis is, as a rule, easy. If diabetes is known to exist, of course the matter is still simpler. The eruption has a markedly neoplastic appearance. Xanthoma diabeticorum is distinguished from ordinary xanthoma by the following characteristics:

1. The rapid evolution and involution of the eruption.

In ordinary xanthoma, the evolution is slow and involution seldom occurs, the lesions usually persisting.

2. Firmness and hardness of the papules.

3. Absence of patches or striæ.

4. Difference in color. Often a distinct reddish tinge, with sometimes yellow apices.

5. Subjective symptoms. Itching, tingling, or tenderness present in xanthoma diabeticorum absent in ordinary xanthoma.

6. Localization. Seats of predilection upon elbows, knees, loins, and buttocks. In ordinary xanthoma the region *par excellence* involved is the upper eyelid.

7. Hair follicles involved.

8. Existence of glycosuria.

The *histological pathology* of the subject has been carefully studied by Morris and Clarke, Crocker and Robinson.

It is pretty generally accepted that xanthoma diabeticorum is an inflammatory affection. Its evolution and involution point indubitably toward this. Large endothelial and giant cells are seen in chronic inflammations, and the fatty degeneration which occurs has its analogue in atheroma of arteries.

As far as the ordinary xanthoma is concerned, this is by no means an established fact. Touton strongly advocates the neoplastic character of xanthoma; others believe it to be inflammatory. There is overgrowth of connective tissue with the formation of groups of large epithelioid fatty cells probably derived from the connective-tissue corpuscles. The rete mucosum contains pigment. Crocker says, "The diseased process in xanthoma diabeticorum appears to be anatomically of the same nature as in ordinary xanthoma, but with more inflammatory phenomena and less connective-tissue growth." Xanthoma cells are found in both forms, but in the diabetic variety to a lesser extent.

In a joint article written by Malcolm Morris and Jackson Clarke, the latter quotes Pye-Smith as saying that "xanthelasma is essentially a chronic, deep dermatitis with early fatty degeneration, the yellowish color depending upon the presence of innumerable fatty granules in the tissues. In the nodules there is also present a dense fibrous tissue, and even in the plane variety a few ill-formed cells have been discovered. The minute structure of xanthelasma is identical with that of atheroma of an artery." Clarke adds that in suitably prepared sections of atheroma the same large "xanthoma cells" are found.

Bacteriological examinations have all been negative.

According to my knowledge, the following is the first case of xanthoma diabeticorum observed in Philadelphia and the third in America:

C. K., aged thirty-four, saloon proprietor, native of Germany. Father died of traumatic myelitis at the age of fifty-three; mother, of unknown cause, at the age of seventy-two. Patient has had measles, varicella, etc., but has since adolescence enjoyed unusual health. Has been a moderate consumer of alcohol and tobacco. Was accepted by a life-insurance company in June, 1893, as a particularly good risk. Stature, five feet five inches. Weight, one hundred and eighty pounds. Two years ago the patient weighed two hundred and eight pounds, but has gradually come down to his present weight. The patient in September, 1893, consulted Dr. Jurist, through whose kindness I have been enabled to see and report the case. At this time the patient was feeling very well, and merely saw his physician because of his skin affection. The character of the eruption, however, at once directed attention to the urine, the examination of which, with the ordinary tests, showed an abundance of glucose. A very small amount of albu-

min was also present, but no casts. Upon close interrogation the following facts were elicited: The patient's thirst was a little more marked than usual, and the amount of urine voided greater. An accurate observation of the amount passed in twenty-four hours showed it to be sixty-four ounces. Appetite normal. No appreciable loss of strength.

Six weeks after the onset of the skin manifestations, the eruption presented the following characteristics: The lesions were pinhead to pea sized, obtusely conical, discrete papules of a dull-red to a yellowish-red color, of dense consistence, sharply defined, and elevated above the skin about one sixteenth of an inch. Upon pressure the redness disappeared and the lesions became distinctly yellow. By the aid of a magnifying glass, and in some lesions without, small dilated capillaries were visible. There were no pustular-looking summits, as has been observed in some cases. The whole lesion was of uniform color. The eruption began upon the elbows, then developed rapidly upon the forearm, lumbar and gluteal regions, axillary folds, and neck respectively (see cut). There were approximately seven hundred papules distributed as stated below: About twenty lesions upon the flexor surfaces of the left arm, double that number upon the extensor surface of the left forearm, about one dozen upon the flexor surface of right forearm, and a similar number upon the posterior aspect of the neck. The front of the neck showed about a half dozen lesions. Numerous papules studded scabies-fashion about the anterior and posterior axillary folds. The loins were the seat of a large number of symmetrically arranged lesions, as will be seen in the accompanying cut. Innumerable pin-head papules upon the buttocks and upper anterior aspect of thighs. There were no lesions upon the knees or legs. Two or three typical papules were seated upon the dorsal surface of the left hand, and one upon the palmar aspect over the metacarpo-phalangeal articulation of the thumb. The face and mucous membranes were free. There was one patch upon the elbow which consisted of several coalesced papules. It was three eighths by a quarter of an inch in diameter, with an irregular surface, and elevated almost an eighth of an inch above the level of the skin.

The only subjective symptom present was tenderness, especially marked when the patient rested his arm upon a hard surface. During involution there was slight evanescent itching.

The patient was placed upon an anti-diabetic regimen, and jambul and codeine given. The lesions, which up to this time had been spreading from day to day, were immediately held in check. Not one new papule developed after the institution of the above treatment. In one

month such a marked change had taken place that the large coalesced plaque upon the elbow was barely perceptible to the finger passed over it. The lesions later became flatter and some of them brownish red or even purpuric in color.

Only in this stage could an error of diagnosis have been made, and the disease perhaps confounded with lichen planus. The similarity,



FIG. 1.—Xanthoma diabeticorum.

however, between these two affections was by no means marked. Later still the lesions became dirty yellow, due to the disappearance of the capillary injection. Now scarcely a trace of the eruption remains.

The urine was carefully examined from time to time and the percentage of glucose calculated. One week after the institution of

treatment (at which time the eruption began to show distinct retrogressive changes) the sugar was reduced to two per cent. About five weeks later an indiscretion in the diet sent the sugar up to 3.2 per cent. At this time the lesions remained stationary for about a week. Albumin was present upon each examination, but careful search failed to reveal the presence of casts.

Microscopic Examination.—A well-marked papule was excised at the acme of the eruption. Owing to the fact that this was hardened

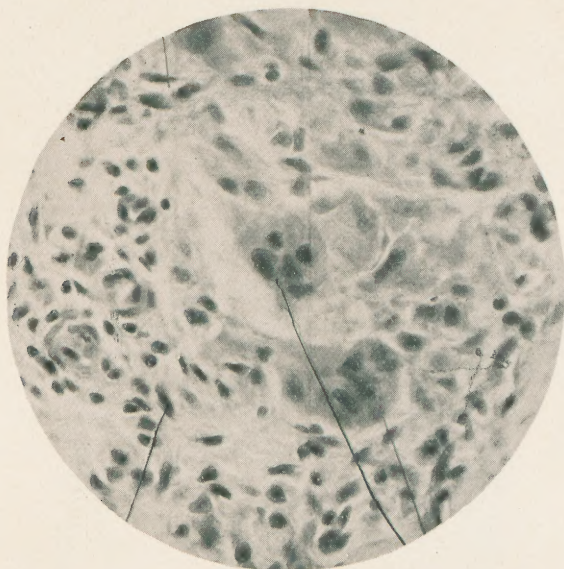


FIG. 2.—The line on the left points to cells derived from the connective tissue corpuscles. Those on the right to the so-called "xanthoma giant cells." Zeiss, DD. $\times 500$.

in alcohol, no fat could be detected later, either in the cells or the interfascicular structures. There was visible under low power a dense but sharply circumscribed cell infiltration involving the papillary bodies and extending deep into the corium. While a large number of these cells were leucocytes, there were also very many connective-tissue cells, spindle or fusiform cells that took the stain (hæmatoxylin) but poorly. The papillary vessels in the vicinity of the infiltration were dilated and the cell walls showed signs of proliferation. A few large endothelial cells were seen lying loose in the lumina of these vessels. Scattered here and there were groups of large epithelioid cells, many of them multinucleated. These are the so-called xanthomic giant cells. They were not present in all the sections, but are quite distinctly shown

in the accompanying cut (Fig. 2). Quite a number of them are grouped about an area just beginning to undergo retrogressive changes. Some of the sweat glands show marked cell infiltration, many of the acini being completely obliterated. A few of the sudoriferous ducts are surrounded by round cells. The papule was in the immediate vicinity of a hair follicle whose root sheaths did not escape the general cell invasion.

I wish here to express my cordial thanks to Dr. Milton B. Hartzell for his kindness in photographing the microscopic preparation.

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